SUNTEXT REVIEWS

SunText Review of Case Reports & Images
ISSN: 2766-4589

Open Access
Case Report
Volume 5:2

Is the Anterior Approach Relevant for Presacral Neurofibromas?

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Received date: 10 February 2024; **Accepted date:** 12 February 2024; **Published date:** 18 February 2024

Citation: Bahrou N, EL Hamzaoui J, Ed-dahby O, El Abidine Khedid YZ, Echarrab E, Elounani M, et al. (2024) Is the Anterior Approach Relevant for Presacral Neurofibromas?. SunText Rev Case Rep Image 5(2): 125.

DOI: https://doi.org/10.51737/2766-4589.2024.125

Abstract

Presacral neurofibroma is a rare benign tumour that originates from nerve sheaths, these masses pose real challenges in both diagnosis and treatment due to its infrequency and unique location. In our report, we discuss the case of a 36-year-old woman experiencing lumbosciatic pain, whose presacral neurofibroma was identified through preoperative imaging and confirmed via histopathological analysis. Surgical removal of the tumour was successful, resulting in positive postoperative outcomes. This case emphasizes the importance of utilizing preoperative imaging techniques and surgical expertise to effectively manage presacral neurofibromas. Furthermore, we provide a comprehensive overview of existing literature, discussing various diagnostic methods, treatment options, and potential complications associated with this uncommon tumour.

Keywords: Neurofibroma; Presacral; Benign tumor; Case report; General surgery

Introduction

Neurofibromas are non-encapsulated neurogenic tumors originating from nerve sheaths, they account for 5% of benign tumors in soft tissues. These ectodermal neoplasms are characterized by their exceptional rarity, both in terms of occurrence and specific anatomical localization. Predominantly found in the thoracic region, followed by the cervical and lumbar regions, merely 1 to 5% of neurofibromas manifest in the presacral region [1]. The intricacies of this anatomical region necessitate a comprehensive, multidisciplinary approach for optimal management, particularly within the framework of a university hospital [2,3]. Typically exhibiting paucisymptomatic or asymptomatic characteristics, these masses often present diagnostic challenges with inconclusive clinical examinations. Diagnosis frequently occurs at an advanced stage of development. Recognizing the anatomical intricacies, preoperative imaging assumes paramount significance in establishing a definitive diagnosis and formulating an effective therapeutic strategy [2,3]. We herein present a case study detailing a pelvic mass identified as a presacral neurofibroma.

Case Report

A 36-year-old female patient with no notable medical history presented with progressively worsening lumbosciatic pain. Lumbar MRI revealed a small paramedian disc herniation at L5-S1, accompanied by a hypodense tissue enlargement of the L5 root extending over 65mm (Figure 1,2). Subsequently, the patient underwent posterior surgery for mass reduction, and histopathological analysis confirmed a neurofibroma through immunohistochemical examination. A follow-up angioscan, conducted 10 months later, identified a lesion at the level of the first sacral foramen measuring 35*38 mm, compressing the psoas muscle, iliac artery, and anterior and posterior trunks (Figure 3). Opting for an anterior approach, a midline incision was made, revealing a small mass below the external iliac pedicle. Following peritoneal opening and dissection of vital structures (urethra, external and internal iliac pedicle) (Figure4), the encapsulated mass was visualized and completely resected, ensuring hemostasis (Figure 5). Immediate postoperative outcomes revealed minimal monoparesis of the right lower limb,

spontaneously regressing. The patient was discharged on the third postoperative day. Histopathological examination revealed a fusocellular proliferation characterized by interwoven bundles of spindle cells with dark and undulating nuclei, interspersed with collagen bands and devoid of mitotic activity (Figures 6,7).

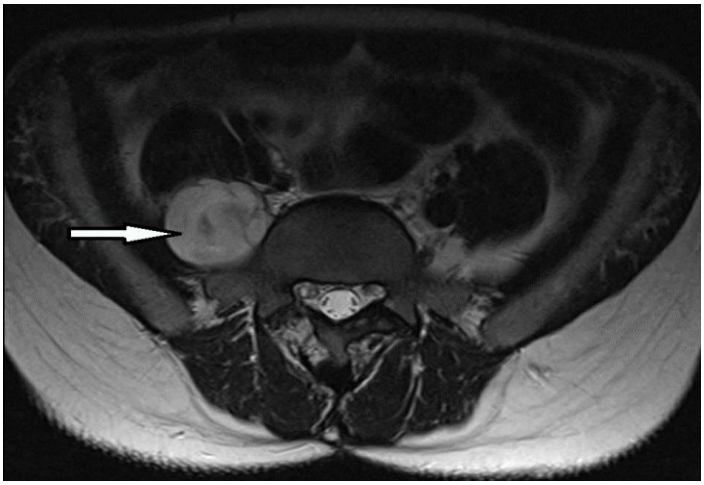


Figure 1: *MRI Axial view.*

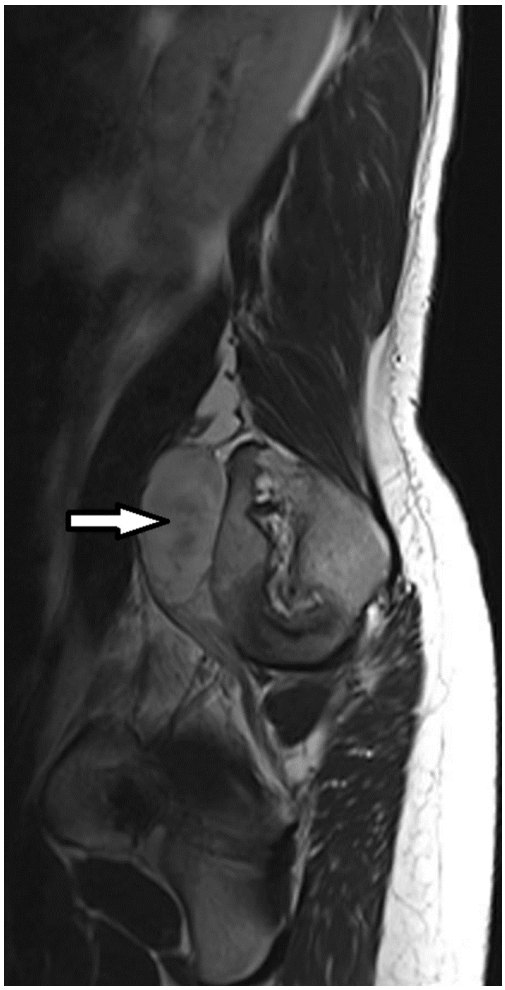


Figure 2: *MRI sagittal view.*

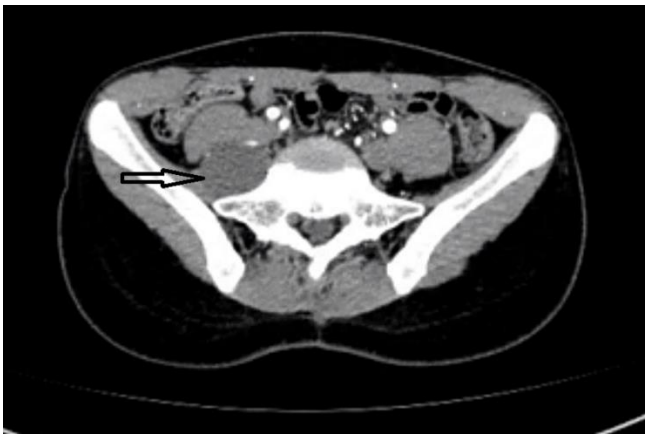


Figure 3: *Angioscan axial view.*

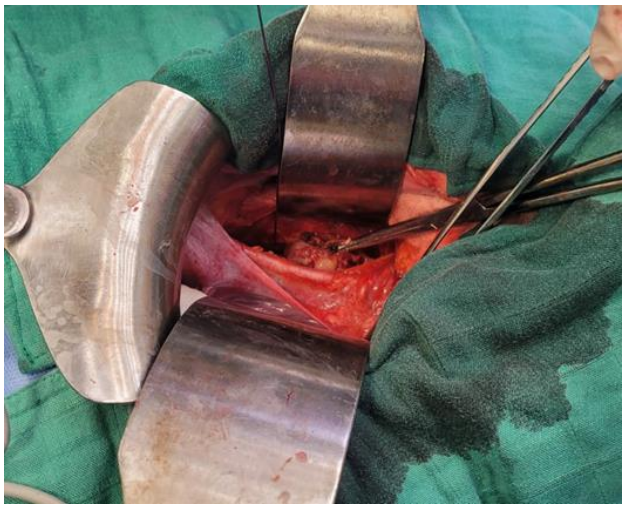
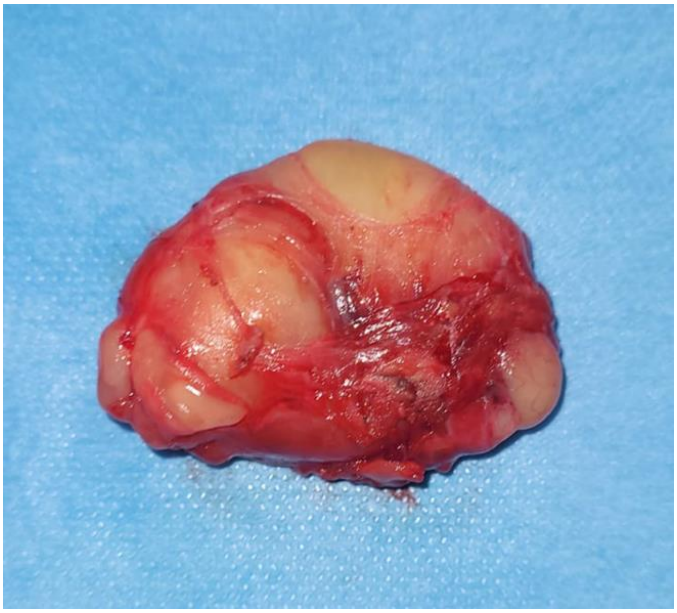


Figure 4: *Peroperative view of the mass adjacent to the external iliac pedicle.*



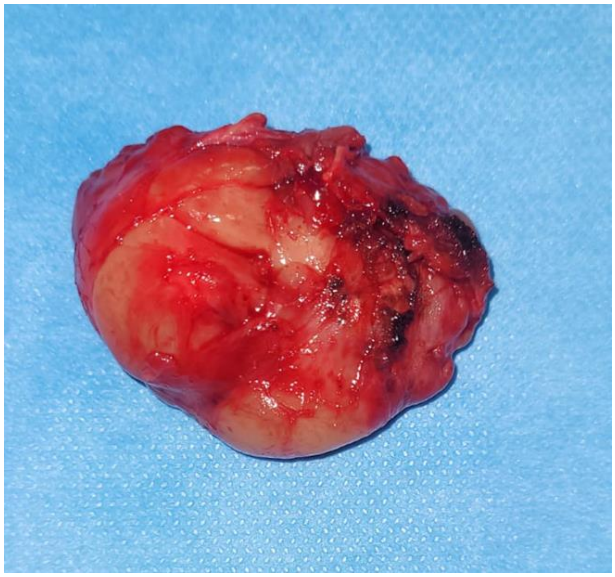


Figure 5: *Peroperative view of the surgical specimen.*

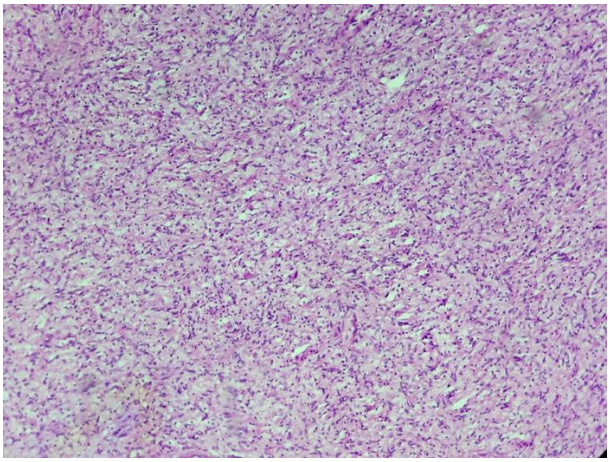


Figure 6: *Neurofibroma: fusocellular proliferation HEX10.*

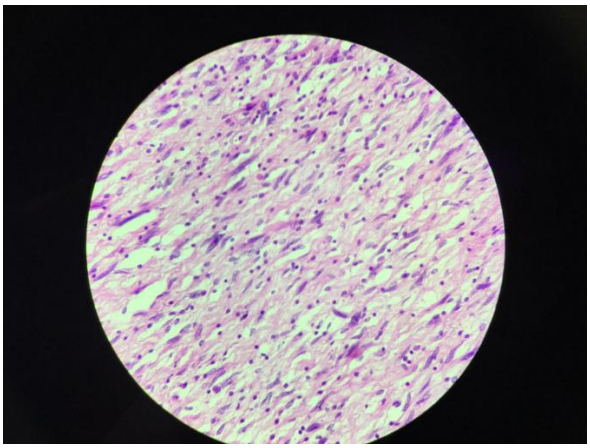


Figure 7: *Presacral neurofibroma : fusocellular proliferation characterized by interwoven bundles of spindle cells with dark and undulating nuclei, interspersed with collagen bands HEX40.*

Immunohistochemical staining with anti-PS 100 antibodies exhibited diffuse marking on the tumour cells, indicative of a neurofibroma diagnosis. Short and long-term follow-up through repeated clinical examinations showed no signs of recurrence or complications.

Discussion

Retroperitoneal masses encompass a variety of entities, ranging from neural tumors like schwannomas and neurofibromas, to fatty tissue lesions such as lipomas and liposarcomas, along with desmoids, lymphomas, and vascular tumors like hemangiopericytomas and angiosarcomas [4]. Neurofibromas, which are rare ectodermal tumors, exceptionally involve the retroperitoneum. They may appear as solitary growths or, more commonly, as multiple lesions associated with conditions like neurofibromatosis type 1 (NF1) or Von Recklinghausen's disease, which is the case with our patient [5]. These tumors tend to grow slowly, extending into the retroperitoneum, presacral space, and soft tissues. In most cases, they do not degenerate, except in 4 to 11% of instances linked to NF1 [2]. Our case is unique due to its rare presacral localization [6]. These tumors often manifest at a young age [1] and remain largely asymptomatic for an extended duration until reaching considerable sizes [7]. This growth can lead to symptoms such as a sensation of heaviness, digestive issues, and urinary symptoms, including hematuria, dysuria, pollakiuria, and even renal colic. Additionally, these masses can be complicated by a massive retroperitoneal hematoma. Preoperative imaging is essential to establish the diagnosis. During ultrasound examination, neurofibromas appear as well-defined masses, hypoechoic and homogeneous, with posterior enhancement [8]. Computed tomography (CT) assesses the tumor's extension to adjacent structures, particularly the sacral plexus, but may not always differentiate between giant neurofibromas and neurofibrosarcomas. Magnetic resonance imaging (MRI) stands out as the most effective modality, providing precise details about the tumor's location and its relationships with neighboring structures. In T1-weighted images, its intensity is slightly higher than that of muscle, appearing hyperintense in T2. Following gadolinium injection, contrast enhancement is heterogeneous, with a slight central hypointensity in T1 [9].

Performing a parietal biopsy is not recommended by some authors, as it often yields inconclusive results and poses risks of hemorrhage, infection, and tumor dissemination [10]. Managing these types of tumors requires a multidisciplinary team comprising visceral, orthopedic, and neurosurgeons [12]. The standard treatment involves a carcinological surgical excision with negative margins, emphasizing the preservation of adjacent vascular, nervous, visceral, and bone structures. The choice of the surgical approach depends on the degree of intra-pelvic and intra-

sacral development. Specifically, an anterior approach is favored for tumors with a significant presacral component, while a posterior approach is suitable for intra-sacral or intra-dural components. Occasionally, a combined sacral and abdominal approach may be employed, as seen in our patient's case [12]. This excision demands a certain level of surgical expertise to avoid severe complications, including injuries to the presacral venous plexus leading to hemorrhage, or damage to the rectum or sacral nerves [13]. However, if the tumor invades adjacent structures, the excision may be extended to achieve negative margins, albeit at the cost of neurosensory deficits [11]. In histopathology, a neurofibroma may present polymorphic cells, including Schwann cells, perineural cells, and fibroblasts contained within a mucopolysaccharide matrix. It infiltrates between the nerve fascicles along its trajectory, making its resection potentially difficult and risky. Immunohistochemical analysis reveals that neurofibroma cells exhibit weak reactivity with the S-100 protein, in contrast to schwannomas, where proliferation exclusively consists of Schwann cells, and positive S-100 immunostaining occurs within the endoneurium of a nerve fascicle [11]. As for radiotherapy, it is generally discouraged for benign tumours due to its potential oncogenic risks [12].

Conclusion

Presacral neurofibroma is a rare benign tumour that often progresses asymptomatically until it reaches significant sizes. Preoperative imaging aids in diagnosing the condition, mapping the lesion, and planning a surgical strategy. The preferred treatment is complete surgical excision, requiring surgical expertise to prevent major complications.

Acknowledgments

None

Data Availability Statement

The data that support the findings of this study are available from the corresponding author upon reasonable request.

Consent of publication

The patient consented to use her data for scientific study and publication purposes.

Funding

The authors have no source of funding or financial support except themselves.

Competing interests

The authors have no conflicts of interest and source of funding. The subject of study had no commercial interest, no financial or material support.

Ethics statement

All Authors declare no conflict of interest.

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